THE DEVELOPMENT OF THE EXTERNAL EAR

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FISTULA AURIS CONGENITA

The little pits or fistulae, known by a variety of names, such as helical fistulae, pre-auricular fistulae, etc., which occur in the neighbourhood of the tragus and crus helicis, are probably present as fairly frequent anomalies in all races. In the African Negro of Nyasaland, Stannus (11) found them in 5·2 per cent. of females and 3·6 per cent. of males. Congdon has recently described a large series in Siamese and other races in Bankok (2). They occur (with a frequency of about 2 per 1000, according to Eyle) in the white races all over the world, and they have long been known to be frequent among the Chinese.

Depressions may occur at various sites upon the auricle, but the present authors agree with Ballantyne (1) that the term fistula auris congenita should be restricted to those pits and fistulae that are situated on, or anterior to, the crus helicis. The pit, and the fistulous track that sometimes leads from it, are definitely congenital: at times they are bilateral, but more usually unilateral. In Stannus's African series the occurrence upon both sides was 0·77 per cent., upon the right side alone 2·08 per cent. and upon the left side alone 1·69 per cent. A definite hereditary history is present in most cases. An example was shown at the Royal Society of Medicine by Dr Prichard (quoted by Stannus) in an infant having bilateral fistulae and whose mother, four siblings, maternal grandmother and two great aunts all exhibited the same anomaly.

As to the commonest site of occurrence, it is probable that some clarity of thought has been lost by the recording of small puckers in varying parts of the auricle and regarding them as true pre-auricular fistulae. Stannus has classified the sites in which his 292 observed fistulae occurred into five positions, and these accord so exactly with our own findings that they are here combined into one figure (see fig. 1 A). The line upon which these fistulae occur may be defined as a curve passing from the temple to the anterior margin of the crus helicis and so to the interspace between the crus helicis and the upper part of the tragus; thence into the concha to the external auditory meatus and out again between the lower part of the tragus and the anti-tragus to the skin at the junction of the concha and the skin of the cheek.

Although these fistulae are undoubtedly of congenital origin, there has always been a difficulty in accounting for their formation. Stannus assumed them to be "remains of the first branchial cleft," and such an assumption has

been made by other observers. But it is obvious that their site of election does not occur along the line of closure of the first depression, as that is pictured in the generally accepted account of the development of the external ear. In discussing their origin, Ballantyne came to the conclusion that "there are two difficulties (at least) in the way of a full and free acceptance of this view" (that they are derivatives of the first depression). "The first is the freedom of the middle ear from participation in the deformity, and the other is the position of the fistulae."

Stammers (10) concludes his account of two cases and a review of the literature by saying: "these fistulae are not remnants of the first branchial cleft,

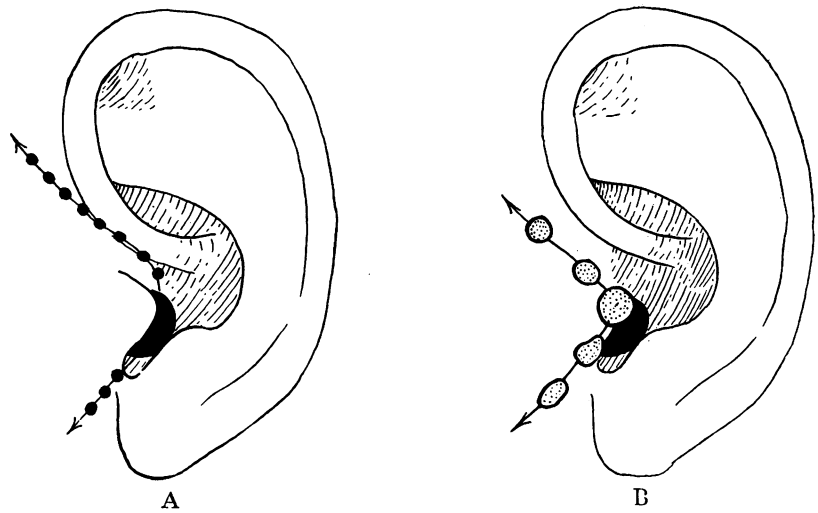

Fig. 1. A. The distribution of pre-auricular fistulae. B. The distribution of pre-auricular appendages.

but result from aberrent coalescence of the six tubercles which are destined to form the pinna." Congdon, in the most recent paper on the subject, considers them to arise from the intertubercular grooves and postulates that shifting of the skin, during the processes of growth, accounts for their general disposition.

There are many difficulties in the way of assuming that these pits and fistulous tracks are caused by faulty union of the so-called auricular tubercles or hillocks of the embryo. In the first place, their distribution by no means appears to follow the lines of union of the several hillocks, as these were depicted by His (7) and later by Streeter (12): and in the second, the hillocks themselves, even as pictured by these investigators, are of such low elevation that it is hardly possible to conceive of a pit or a fistula being formed at their sites of junction.

PRE-AURICULAR APPENDAGES

One point of considerable interest lies in the fact that pre-auricular fistulae are at times complicated by the presence of accessory auricular appendages, as in the cases recorded by Ballantyne and by Congdon, and it is therefore possible that some light may be thrown on their origin from an examination of these appendages.

Pre-auricular appendages (accessory auricles, supernumerary tragus, etc.) are of fairly common occurrence. By Warner they were reported 33 times in the routine examination of 50,000 children. It must not be forgotten that these little skin tags are often removed for cosmetic purposes; but, if bifid tragus be included under this heading, they would appear to have a far higher frequency in the Chinese. Six Chinese foetuses, out of our series of 400 at the Peiping Union Medical College, showed this condition either unilaterally or bilaterally.

The site of pre-auricular appendages appears to be very constant: from the tragus as a centre they may extend upwards and forwards towards the temporal region, between the upper part of the tragus and the crus helicis; or downwards towards the cheek, between the lower part of the tragus and the anti-tragus. It is therefore clear that the line of election of pre-auricular appendages lies just in front of the line of election of pre-auricular fistulae (see fig. 1B). It would certainly seem that this general agreement of the sites of these two anomalous conditions of the auricle argued for a common and normal developmental process underlying them both.

THE DEVELOPMENT OF THE EXTERNAL EAR

Before His published his classical account of the development of the external ear, the descriptions of the process available in the literature were very meagre. Warton Jones's account (8) is difficult to follow and no distinctions are made between the portions of the ear arising from the mandibular arch and those developing from the hyoid. Allen Thomson's work (13) led to some amplification of the descriptive details, and his statement that "the pinna is gradually developed on the posterior margin of the first branchial cleft" remained as the accepted account of the process for thirty years and through successive editions of Quain's *Anatomy*. It would seem that, previous to the description published by His, the pinna was regarded as a hyoid derivative.

According to His's researches, the pinna was developed from three tubercles or hillocks situated on the mandibular arch and three on the hyoid arch. The after history of these hillocks and the respective shares that the mandibular and hyoid hillocks have in the formation of the definitive auricle have been rather variably estimated by a series of later investigators—Gradenigo, Schwalbe, Baum and Dobers, Henneberg, Streeter and others.

The main point of interest, from a morphological point of view, is how much of the definitive external ear is mandibular and how much hyoid in origin.

According to His, the mandibular contribution embraces the tragus, the crus helicis, and the helix itself as far as the tuberculum auriculae. Other investigators have differed from His mainly in defining other limits upon the helix as the termination of the mandibular element. It is at once apparent that the precise determination of the site of junction of hillocks 3 and 4 (or the meeting of mandibular and hyoid elements) is a matter of some importance, since this point must mark the upper limit of the first pharyngeal depression.

If the current account of the development of the external ear be correct, we must assume that the line of the first pharyngeal depression passes upwards and backwards from the incisura intertragica and across the meatus auditorius externus to the region of the tuberculum auriculae (see fig. 2).

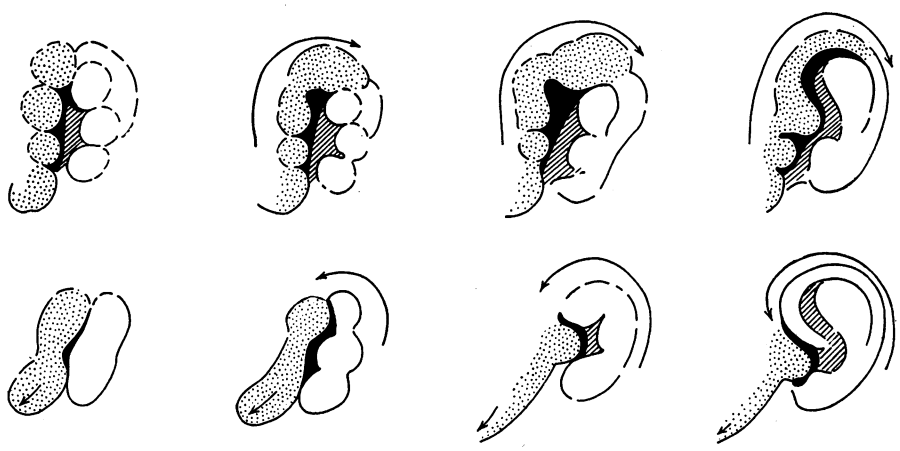

Fig. 2. The development of the external ear. The fate of the mandibular and hyoid elements as depicted by His, etc. (upper series), and as determined in the present investigation (lower series).

It would seem that such a disposition of the first depression is unlikely, since the crus helicis lies definitely behind the meatus or on the hyoid side of the depression. By the classical description a migration of the upper two mandibular tubercles is demanded, the migration taking place in a dorsad and caudad direction so that a considerable portion of the pinna on the hyoid aspect of the meatus is postulated as having shifted from the mandibular side.

In order to test the reality of this migration of the mandibular portion of the pinna, we re-examined the series of embryos in the collection of the Peiping Union Medical College. Our initial difficulty in studying the developing auricle in these well-preserved, formalin-hardened, embryos was the lack of any confidence we felt in identifying the six hillocks that constitute the fundamental features in the classical accounts of the process. This difficulty applied more particularly to the mandibular arch; for upon the hyoid arch it was often possible to detect slight elevations in the caudal boundary of the first depression. Feeling that this inability to detect the hillocks with any real

conviction might be due to our own faulty observation, we gave the series of embryos to the departmental Chinese artist, with instructions to produce enlarged drawings of exactly what he could see in the neck region. The latter's drawings of a series of embryos ranging from 12 to 24 mm. coincided exactly with our own observations and differed widely from the classical figures given by His, but they very strikingly resembled those which illustrate Frazer's work (see especially *J. Anat.* vol. LXI, pp. 132–43). It is possible that this discrepancy is accounted for by the method of preservation, and that shrinkage in spirit may render these hillocks more apparent, or may even determine their presence. It would appear that others have had difficulty in identifying the hillocks and in following their transitions, for Keibel (9) states, "I have not been able to get a clear picture from the investigation of the human embryos at my disposal, but it seems certain that the tragus is developed from the first auricular hillock and the antitragus from the sixth, and that the auricular lobe is a later formation that has nothing to do with the hillocks."

It is apparent, from the first stage at which the rudiments of the auricle are visible, and throughout the whole of the subsequent stages, that the elevation on the hyoid arch is far more extensive and far more prominent than is that on the mandibular arch. It is also apparent that the mandibular portion is growing forwards (ventrad) on the anterior aspect of the first depression as the prominence of the chin region begins to develop upon the growing jaw. Such a movement would seem to indicate that, if there be any translation of the hillocks from one side of the depression to the other (as is postulated on the classical description), it would be evidenced by the higher hyoid hillocks (Nos. 4 and 5 of His) passing round the upper margin of the depression, rather than in a migration of the upper mandibular hillocks (Nos. 2 and 3 of His) to the hyoid aspect of the depression. The process of development, as it appears to be demonstrated in our series of embryos, appears to consist in the formation of a raised margin upon the hyoid boundary of the first depression. This elevation presents a somewhat tuberculated appearance at certain stages and these tubercles might perhaps justify the name of hillocks: but we agree with Keibel that "the investigation of the further history of these structures is unusually difficult." The difficulty appears to arise from the inconstancy of the hillocks themselves. The hyoid elevation differentiates into an auricular fold before any structural landmarks are apparent upon the mandibular elevation. The hyoid auricular fold extends beyond the most dorsal limit of the first pharyngeal depression, and soon encircles the upper end of the depression. This movement appears to be consequent upon the forward growth of the mandible. The mandibular boundary of the depression now shows a definite elevation, which in some specimens shows evidence of partial subdivision into tubercles. At the junction of the hyoid auricular fold and the upper part of the mandibular elevation an inflection takes place and the hyoid portion curves down so that it sweeps in and forms the crus helicis, which however never succeeds in passing to the mandibular aspect of the depression, but distorts the upper part

of the depression in its growth. In this way the mandibular elevation is depressed and persists as the definitive tragus. It would seem from the figures given by Frazer ((4) p. 139, fig. 7) that the process here described is exactly the same as that seen by him when making his drawings, for the whole of the auricle, with the exception of the tragus, appears to be formed from the hyoid elevation.

In fig. 2 (lower series) we have endeavoured to show the processes as we conceive them to take place contrasted with those invoked in the classical description. It may appear to be a retrograde step, but we conclude that Allen Thomson's observation ("the pinna is gradually developed on the posterior margin of the first branchial cleft") made in 1844 is not far from the truth. The tragus appears to be the only mandibular derivative of the pinna and the line of the first pharyngeal depression appears to coincide with the line of election of pre-auricular fistulae.

THE EXTERNAL EAR IN CASES OF ARRESTED DEVELOPMENT OF THE MANDIBULAR ARCH

A very large number of cases of failure of the mandibular arch to be fully developed are recorded in the literature. Failure of development may express itself over a wide range of malformations, ranging from trivial degrees of micrognathia to complete agnathia. These anomalies occur in lower animals as well as in man, and the peculiar frequency of an agnathic condition in lambs is well known to sheep breeders. Although associated malformations of the pinna have been reported in certain human cases, they are by no means a necessary concomitant of the primary malformation, nor indeed are they its usual accompaniment.

The typical condition seen in cases of agnathia is for the whole of the pinna to be developed behind the meatus auditorius externus;—but the tragus is entirely unrepresented. A very complete case of agnathia is that described by Hannover 50 years ago (6). Two malformed auditory ossicles and three minute spicules of bone alone represented the mandibular arch; and yet only the tragus was missing from the external ear. Hannover's original illustration is reproduced here as fig. 3. Another case, which duplicates that of Hannover in almost every detail, is that described by Graham (5) and in this case also the external ear was complete with the exception of the tragus. In a specimen examined for the purposes of the present inquiry (Melb. Univ. Path. Mus. No. 62A) the region of the first arch had already been dissected and all traces of it are said, on the label, to have been absent. The external ear of this specimen is illustrated at fig. 4. The tragus is absent, but with that exception the external ear is well developed, save that the lobule is not well defined and is adherent. These examples are typical of the condition, and it would therefore seem to be a definite fact that absence, or almost total absence of the first pharyngeal arch, involves no more deformity of the external ear than the lack

of development of the tragus. The helix being well developed in cases of agnathia, would certainly seem to indicate that it was not formed from the mandibular arch.

THE NERVE SUPPLY OF THE EXTERNAL EAR

There is a marked discrepancy between the nerve supply of the pinna as determined by anatomists and that commonly recognised by the clinician. The anterior auricular branches of the auriculo-temporal nerve are accepted, in the standard anatomical descriptions, as supplying the front of the upper part of the auricle. The area of supply by the trigeminus is usually depicted as including the whole of the upper part of the pinna down to about its mid-point on the posterior margin, sometimes including the tragus (Quain) and sometimes

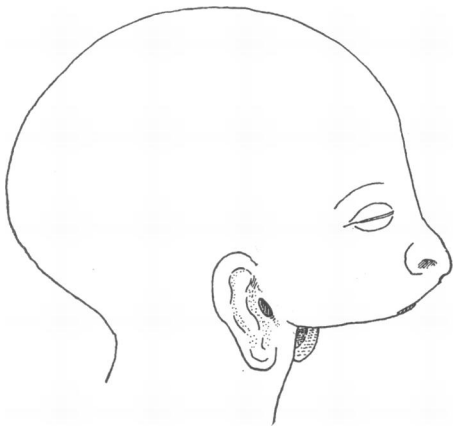

Fig. 3. The external ear in Agnathia. Hannover's case, 1884.

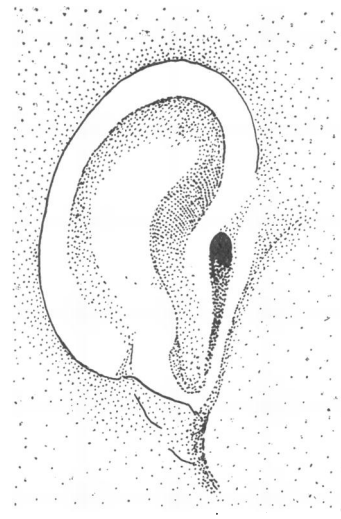

Fig. 4. The external ear in Agnathia. Melbourne University Path. Mus. No. 62 A.

omitting it (Gray) in front (see fig. 5 A). Nevertheless, although at the present day there seems to be a general agreement among anatomists that the auriculotemporal nerve supplies a large part of the pinna, this has not always been the accepted opinion. Macalister, in 1889, described the auricular branch as being "lost on the skin of the tragus and vicinity of the intertragic notch"—a description almost identical with that of Holden (1861), Knox (1853), Ledwick (1852) and other nineteenth century anatomists. There is no doubt that this earlier description is adequate for a great many cases; in others, branches, apparently from the auriculotemporal, may be traced to the upper part of the pinna. It is, of course, likely that the actual distribution of the cutaneous nerves supplied by this branch of the trigeminus varies within fairly wide limits: but it is also likely that, in many cases, the branches identified in gross

dissection are actually those of the nervus facialis that are running with the trigeminus. The great interchange of fibres between the trigeminus and the facialis has taken place before the anterior auricular branches are given off from the former. In some cases, even by gross methods, it can be demonstrated that the auricular branch proceeding to the top of the pinna is destined for the supply of m. helicis major. In lower Mammals (such as *Macropus*) it is obvious that, whilst the auricular branches of the facialis pass up to supply the muscles of the large pinna, the auricular branch of the trigeminus goes no farther than

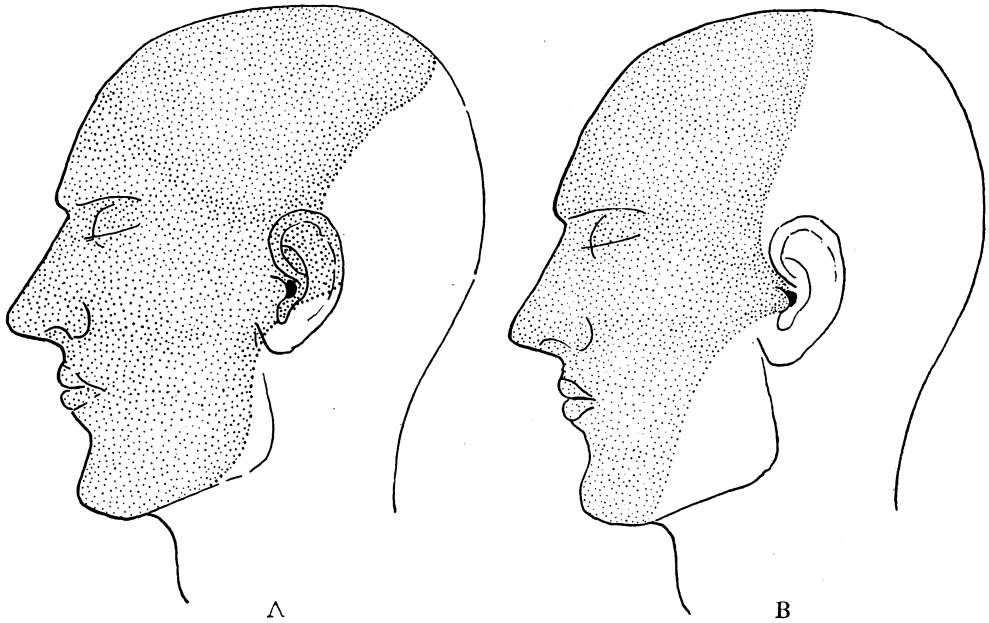

Fig. 5. The distribution of the trigeminus to the ear (A) according to most anatomists (after Thane); (B) according to most clinicians (after Foerster, Head and Cushing).

to the front of the external auditory meatus. It is, therefore, highly probable that the true supply of the auricular branch of the trigeminus passes no farther on to the pinna than the tragus and the anterior aspect of the external auditory meatus as Macalister affirmed.

In neuritis of the trigeminus the pinna itself is not painful; and, conversely, after removal of the Gasserian ganglion or section of the third division of the trigeminus the anaesthetic area does not invade the pinna. There is now very definite agreement among clinicians that the trigeminus supply is limited to the tragus and to the anterior part of the external auditory meatus (see fig. 5B). On the other hand, section of the great auricular nerve produces anaesthesia of the whole of the pinna with the exception of the anterior part of the external auditory meatus and the tragus.

CONCLUSION

It appears from the study of the development, anomalies, and nerve supply of the external ear, that the pinna is a hyoid derivative and that the mandibular contribution is represented only by the tragus and the anterior part of the external auditory meatus. It is, therefore, evident that pre-auricular fistulae and pre-auricular appendages do, in reality, lie along the line of the first pharyngeal depression.

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